

Modern Concepts of Cardiovascular Disease

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THE CLINICAL ASPECTS OF CONGENITAL CARDIAC DISEASE

Part I

GENERAL CONSIDERATIONS AND HISTORICAL SURVEY

During the past thirty years, this subject, formerly of little more than academic interest, has come into general recognition as a special department of clinical cardiology of great practical importance and high scientific value. Many factors have contributed to the elucidation of this hitherto obscure field. Already, in the earlier part of the last century, ground had been broken for future exact investigation by the clear delineation of the special structural changes and clinical features presented by these defects, in many careful reports published from reliable sources in the Transactions of the Pathological Society of London and elsewhere as well as in the early surveys by Farre (1814), Chevers (1846), and others, culminating in Peacock's classic treatise "*On Malformations of the Human Heart*" (1858 and 1866), and Rokitsansky's epoch-making contribution "*Die Defekte der Scheidewände des Herzens*" (1875). In the latter work the pathogenesis of cardiovascular septal defects complicated by other grave anomalies was made for the first time the subject of an intensive systematic study with illuminating and far-reaching results, and the promulgation of a brilliant hypothesis that has found at least partial confirmation in the researches of later observers. We owe it indeed to his genius as well as to the investigations into comparative embryology by His (1886), Born (1889), Röse (1890) and Greil (1903) and the phylogenetic studies upon the explanation of cardiac anomalies based upon the above by Sir Arthur Keith, Jane Robertson, Waterston and Spitzer, that a rational theory of the ontogenesis of these hitherto obscure conditions is at last supplied.

Other advances which have conduced to our clearer understanding of this subject are incidental to the immense progress made in recent years in all departments of modern medicine, particularly

as regards laboratory methods and diagnostic procedures. Such are, the studies by Haldane and his co-workers into the biochemistry of the cardio-respiratory mechanism and the blood changes in congenital cyanosis; the calculation of the venous-arterial shunt in these cases by Parkes Weber and Dorner (1911), and more recently by Weiss and Löwbeer (1924), Lundsgaard and Van Slyke, and others; the clarification of this entire subject of cyanosis by the last named authorities in their important monograph in *Medicine*, 1923; the fundamental studies carried on by Krogh and his associates with the aid of the capillary microscope (1929); the formulation by Redisch and Rösler of their theory on the capillary origin of cyanosis; the studies by Lewis and Grant on bicuspid aortic valve and the frequent incidence of infective processes in this anomaly; our growing knowledge of the fluoroscopic findings in congenital heart disease and their application in the differential diagnosis of acquired lesions; and the popularization of this subject by the emphasis laid upon its clinical aspects by Dr. Paul D. White in his recent textbook on Heart Disease.

Finally, careful consideration of the clinico-pathological aspects of this subject reveals an interesting observation which we believe to be of paramount importance for an intelligent grasp of its clinical values. We refer to the fact that there are to be recognized several distinct types of cardiac anomalies, which differ essentially from each other both in the nature of the underlying structural changes and in their symptomatology and course. As a result, a Clinical Classification has been evolved, based upon the pathological physiology of the circulation as altered by the defect and the special symptoms thereby induced. (Published by Abbott and Dawson with illustrative diagrams of

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the circulation in the International Clinics, 1924). Under such an arrangement the cases of clinical significance may be placed in one of the three following categories:

- I. *Acyanotic Group*. Cases in which there is no abnormal communication between the two circulations, so that no cause for cyanosis exists, but the anomaly is the seat of strain.
- II. *Cyanose Tardive*. Cases in which there is absence of cyanosis except as a transient or terminal feature. These are the localized cardiovascular septal defects, patent ductus and foramen ovale, in which there occurs an arterial-venous shunt from left to right through the defect with possible terminal reversal of flow.
- III. *Cyanotic Group*. Cases presenting constant, progressively increasing cyanosis, due to a permanent venous-arterial shunt, or prolonged deoxygenation in the capillaries, the result of grave structural changes, and giving rise to the familiar symptom-complex of persistently raised oxygen-unsaturation.

The recognition of clinical types as outlined above is not new. Already, in 1857, Bamberger had proposed a classification based upon the presence or absence of cyanosis; and the differentiation of "left-sided" (Parkes Weber) and "right-sided" valvular lesions (Newton Pitt) had the same underlying idea. The most fundamental contribution, however, to our understanding of this subject, was made by Bard and Curtillet (*Revue de Médecine*, 1889) in their differentiation of "cyanose tardive" from the permanent "congenital" form as the special characteristic of defects of the interauricular septum or patent foramen ovale setting in only on the onset of some factor raising the pressure in the right heart or pulmonary circulation. Their article, which ranks as one of the classics of medicine, relates the case of a man aged 54, who had been in robust health until some months before death and suddenly became acutely ill with signs of bronchopneumonia, dyspnoea and onset of deep cyanosis of the face and extremities. Autopsy revealed a patent foramen admitting a pen-handle, dilatation of both auricles but especially the right and extensive lobular pneumonia. The authors explain that in the patient's previously healthy life "the pressure being highest in the left auricle" the anomalous shunt had passed from left to right through the defect, so that no cause for cyanosis had existed; but that, on the advent of his bronchopneumonia a reversal of flow had occurred with development of a "late" cyanosis. They cite several other instances of auricular septal defects in robust, elderly individuals presenting the abrupt onset of "late cyanosis" on the advent of pulmonary disease, and close their account of what they rightly considered a new clinical entity as follows: "Thus we see that besides the early better known forms of cyanosis there exists another, the 'forme tardive.' This occurs at an advanced age among previously robust subjects in whom the presence of a patent foramen has not previously been suspected, and it appears suddenly when an acquired factor, whether pulmonary or cardiac, breaks the normal intracardiac relative pressures in favour

of that in the right auricle; . . . thus permitting the passage of venous blood into the left."

Of much interest also, is the establishment by another group of French observers, of the clinical picture of localized defect of the interventricular septum (*maladie de Roger*) as belonging also in the "cyanose tardive" category (Group II). This author had in 1879 published his clinical observation of a healthy young man presenting the distinctive physical signs of a defect at the base of the interventricular septum. Twelve years later this patient died with terminal cyanosis and the autopsy, reported by Dupré (1891), confirmed Roger's diagnosis. Later, Reiss, (1892), and LeHoux (1902), discussed this with other illustrative cases and presented conclusive evidence that absence of cyanosis until advanced life is the rule in uncomplicated septal defect, and that *cyanose tardive* is a not infrequent termination and constitutes here also a part of the clinical picture.

It is in the light of these brilliant contributions to the nosography of the *cyanose tardive* group and those of Fallot and others to the delineation of the congenital cyanotic cases, combined with the results of the writer's statistical survey of the literature, that the Clinical Classification outlined above was formulated. In our judgment the practical value of such a grouping for the correct evaluation of the clinical significance of any defect is very great. In the outline given in the next issue this order will be followed and its importance stressed.

RELATIVE INCIDENCE

Statistics vary, but indicate that the ratio of cardiac anomalies to deaths from all causes is approximately 1%. The incidence is naturally higher in children. This was well demonstrated by Philpott who found among 7,240 autopsies in Montreal Hospitals 80 cardiac anomalies (1.1%), distributed as follows:

- I. Birth to 28 days, 36 cases in 759 autopsies (4.7%).
- II. 1 month to 2 years, 27 cases in 954 autopsies (2.8%).
- III. Above 2 years, 17 cases in 5,522 autopsies (.3%).

A similar ratio seems to apply to congenital and acquired heart disease. White and Jones found among 2,421 cases of organic heart disease at all ages, 37 of congenital origin (1.5%); while the combined statistics from three large Children's Hospitals showed an incidence of 7.6% (Norris). The relative rarity of congenital heart disease in older subjects does not lessen its importance to the internist who must be alive to all contingencies in order to arrive at a sound differential diagnosis of any cardiac lesion.

(To be continued)

References to all the authorities mentioned will be found listed in the bibliography appended to the writer's monograph in Nelson's Loose-leaf Medicine.

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